

Core Decompression in Avascular Necrosis of the Hip in Sickle-Cell Disease

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Sickle-cell disease (SCD) is the most common cause of avascular necrosis (AVN) of the hip in childhood. It results in significant physical impairment and chronic pain, and often progresses to require hip replacement. Conservative therapy is ineffective. We evaluated whether core decompression can arrest progression of AVN. We performed 13 coring procedures in 10 patients with SCD and AVN. Patients ranged from age 9–21 years at diagnosis (mean, median age, 15 years); five hips were stage I, six hips were stage II, and two hips were stage III. Mean follow-up on these patients was 3.7 years. Efficacy of the procedure was evaluated by clinical improvement in pain, radiographic progression, and need for further surgery. All 5 stage I patients had substantial improvement in pain, and only one showed X-ray progression. Five of the 6 (83%) stage II patients had improvement in pain, and 2 patients progressed on X-ray. Both stage III patients progressed on X-ray, but one was clinically improved. None of the 10 patients has required further surgery. Our results demonstrate that in early AVN, core decompression was beneficial for almost all patients, even with progression on X-ray. Core decompression should be considered in the management of SCD patients with early AVN. © 1996 Wiley-Liss, Inc.

Key words: sickle-cell disease, avascular necrosis, core decompression

INTRODUCTION

Sickle-cell disease (SCD) is the most common cause of avascular necrosis (AVN) of the hip in childhood [1]. The overall prevalence of AVN in SCD is at least 10%, and homozygous SCD patients with concomitant alpha thalassemia are at particular risk, with a prevalence of >20% [2,3]. The risk of developing AVN does not disappear with age, and by age 35 years nearly half of all homozygous SCD patients will have developed AVN. Avascular necrosis frequently progresses to total collapse of the femoral head, necessitating hip replacement [1,4,5]. Unfortunately, sickle-cell disease patients have had particularly poor results with hip arthroplasty, with up to 59% failing within 6 years [6–8]. As the life expectancy of SCD patients increases, the morbidity of AVN will become increasingly important.

Avascular necrosis of the hip can often be detected at an early stage, and there is considerable interest in therapies to halt progression of AVN [9–11]. Unfortunately, conservative management, such as prolonged nonweight-bearing or transfusion therapy, is not effective in preventing progression in most cases [12,13]. Core decompression of the femoral head is an alternative therapy which has

been reported to reduce pain and prevent progression of AVN [4,14,15]. There are, however, no reports of its use in children or solely in patients with SCD. We report the results of core decompression in SCD children afflicted with AVN of the hip.

METHODS

Patients

Ten children with SCD underwent core decompression in 13 hips. All patients are followed regularly at the Northern California Comprehensive Sickle Cell Center and had their diagnosis of SCD confirmed by standard electrophoretic techniques. Eight patients had hemoglobin (Hb) SS, one patient had Hb SC, and one had Hb S β^0 thalassemia. Patients ranged from age 9–21 years (mean, median age, 15 years), and there were 5 males.

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Patients were eligible for core decompression if they met the following criteria: 1) severe pain localized to the hip or inguinal area requiring hospitalization with intravenous narcotics, 2) physical examination findings of pain and limitation on movement of the affected hip, and 3) radiographic or magnetic resonance imaging (MRI) findings diagnostic of AVN. After diagnosis with AVN, bed rest was recommended but not consistently followed. Patients had not received any other treatment for AVN (e.g., hydroxyurea or chronic transfusion) prior to the core decompression procedure.

Diagnosis and Staging

All patients had a radiologic evaluation consisting of anteroposterior (AP) and frog-leg views on plain radiography and MRI after complaining of hip pain consistent with AVN. Conventional criteria were used to establish a diagnosis of AVN on plain radiographs, including sclerosis, increased radiolucency, and changes in femoral head shape [5]. When plain radiographs were normal, the diagnosis of AVN was made using MRI, which demonstrated increased signal intensity in the femoral epiphysis. Sixteen hips in 10 patients were found to be diagnostic of AVN. Six patients had bilateral disease, 3 had right-sided disease, and one had left-sided disease. Core decompression was performed in the 13 symptomatic hips, and these were staged according to the Ficat system [5]:

- Stage I. Normal plain radiograph, abnormal MRI.
- Stage II. Sclerosis and lytic areas on radiography.
- Stage III. Flattening of femoral head.
- Stage IV. Collapse of femoral head, joint space narrowing.

At time of surgery, five hips had stage I disease, six had stage II disease, and two had stage III disease.

Operative Procedure

Patients were admitted the day before the scheduled coring procedure and received intravenous hydration and red blood cell transfusion, following a standard protocol for SCD patients undergoing elective surgery [16]. Core decompression was performed according to a previously described procedure [15]. Briefly, in this procedure a guide pin is inserted into the femoral head from below the trochanteric ridge and directed toward the necrotic area (Fig. 1). Correct placement of the guide pin is established with two-plane imaging, and then an 8-mm hollow coring device is used over the guide pin to remove a core of medullary bone. Intramedullary pressure measurements and biopsies were not regularly performed. Patients were discharged with instructions to complete 2 weeks of strict nonweight-bearing followed by increasing activity as tolerated. No other specific treatments, such as antiinflammatory agents or hydroxyurea, were used postoperatively.

Follow-Up Evaluation

After surgery, patients were seen for a follow-up visit at 2 weeks with the hematology service, and again at 6 weeks postoperatively by the orthopedic service. Thereafter, patients were followed with routine comprehensive health care visits every 6–12 months. Patients were evaluated by three criteria: clinical improvement in pain and activity, radiographic progression, and need for further surgery. Clinical efficacy was scored using a four-point scale assigned on the basis of patient report and physician examination according to the following criteria: 1) patient pain free and no limitation of activities or range of motion on exam, 2) much improvement in pain and minimal limitation in activities or range of motion, 3) little or no change in pain, significant limitation of activities or range of motion, and 4) worse pain, more limitation in activities or range of motion. Radiographic evaluation was made approximately every 6–12 months, or more frequently if the patient developed symptoms suggestive of progression. Radiographic follow-up protocol was for plain radiographs on all patients and, in most cases, MRI as well. The decision for further surgery was based on severity of pain and impairment of patient activity: specifically, continued dependence on narcotics and lack of ambulation (unable to bear weight on affected limb).

RESULTS

Stage I

All five hips had a significant reduction in pain (clinical score = 2), although none was pain-free (Table I). The most common remaining complaint was of vague aching in the joint which did not require regular narcotic use or hospitalization. Limitation in activity and range of motion was not seen in the absence of pain. Four of the 5 patients' disease was stable on radiologic evaluation. One of the 5 patients progressed on plain radiography to stage IV disease over the 18 months following core decompression. Despite progression, this patient reported substantial improvement in pain and mobility. No patient has required further surgery. Mean follow-up for this group is 2 years and 8 months (median, 3 years, 3 months; range, 9 months–3 years, 5 months).

Stage II

Five of the six hips (83%) had significant improvement in pain (clinical score = 2) as with stage I patients, and the remaining patient reported little change. Four of the 6 patients showed no radiographic progression of disease. The remaining 2 patients progressed on radiography, one to stage III and one to stage IV. The patient who progressed to stage III had improvement in pain, but the patient who went on to develop stage IV disease reported no change after core decompression. None of these 6 patients has needed further surgery, and mean follow-up



Fig. 1. Radiographic appearance of avascular necrosis of the hip before and after core decompression. Plain radiograph (lower left) demonstrating stage II avascular necrosis, with mild sclerosis of right femoral head (arrows), and MRI (upper left) with increased, asymmetric uptake in the right femoral head. Right: Same patient 1 year later, after core decompression, with stable disease and prominent core decompression track (arrows).

for this group is 4 years, 2 months (median, 3 years, 2 months; range, 8 months–9 years, 9 months).

Stage III

Both hips in this group progressed on plain radiographs. One had significant clinical improvement despite X-ray progression, and the other was clinically unchanged. Neither patient has needed further surgery.

In analyzing all of the groups together, radiographic progression was not consistently associated with change in symptoms after the coring procedure. Three of the 5

patients (60%) who progressed on plain film had substantial clinical improvement, with reduction of pain and increased mobility. Clinical failure, however, did correlate with radiographic failure with both patients who failed clinically, also showing radiographic progression.

Complications

Surgery, in general, was well-tolerated, and patients recuperated quickly. Postoperatively, one patient developed a wound infection and another developed an acute chest syndrome which did not require transfusion. A third

TABLE I. Results of Core Decompression in Patients With Sickle-Cell Disease

| Patient | Age (years)/Sex | Stage ^a | Follow-up length | Postoperative results ^b | |
|---------|-----------------|--------------------|--------------------|------------------------------------|-------------------|
| | | | | Clinical score | X-ray progression |
| 1 | 14/M | I | 2 years, 3 months | 2 | Yes (stage IV) |
| 2 | 21/F | I | 3 years, 3 months | 2 | No |
| 3 | 17/M | I (R) | 3 years, 5 months | 2 | No |
| | | I (L) | 3 years, 5 months | 2 | No |
| 4 | 9/M | I | 9 months | 2 | No |
| 5 | 17/M | II (R) | 8 years, 9 months | 3 | Yes (stage IV) |
| | | II (L) | 8 years, 9 months | 2 | No |
| 6 | 15/F | II | 3 years, 2 months | 2 | No |
| | | II | 3 years, 2 months | 2 | No |
| 7 | 18/F | II | 8 months | 2 | Yes (stage III) |
| 8 | 13/M | II | 10 months | 2 | No |
| 9 | 13/F | III | 2 years, 9 months | 2 | Yes (stage III) |
| 10 | 16 | III | 10 years, 4 months | 3 | Yes (stage IV) |

^aStage at time of core decompression; R, right; L, left.

^bPostoperative results as of last follow-up visit. Clinical score: 1, no pain or limitation of mobility; 2, substantial decrease in pain and increased mobility; 3, little change in pain or mobility; 4, worse pain or more limitation in activities.

patient with a history of severe auto- and alloimmunization was discharged after 4 days but was readmitted 3 days later for a severe hemolytic transfusion reaction due to receiving incompatible blood. Estimated blood loss was minimal in almost every case and less than 10 cc/kg in all patients. Mean length of hospitalization was 7 days (median, 6.5 days). There were no subtrochanteric fractures and no growth disturbance in the 3 patients with open epiphyses at time of surgery.

DISCUSSION

Avascular necrosis of the hip results from severe ischemia of the medullary bone. Blockage of the osseous circulation results in necrosis and in an increase in intramedullary pressure, which then causes further vascular compromise [5,17]. Without treatment, AVN often progresses to total collapse of the femoral head. Core decompression was originally reported by Ficat and Utheza [18] to prevent progression of AVN when performed early in the disease. In this procedure a core of bone is removed from the femoral head, reducing intramedullary pressure and thereby preventing further vascular impingement and allowing for new bone formation [5,15]. Stulberg et al. [14] recently reported a randomized trial comparing core decompression with prolonged nonweight-bearing. They documented that 70% of patients with early AVN treated with core decompression showed improvement, compared with only 20% of those treated with prolonged bed rest. Twenty-eight percent of patients treated with core decompression needed hip replacement, as compared to >50% in those treated nonoperatively. Other series have not had as much success with core decompression, and have reported complications such as subtrochanteric fractures from the procedure [19–21]. It is difficult to compare

these different reports, as there were no separate analyses by etiology of AVN and no standard criteria to define efficacy of the procedure.

We have attempted to perform core decompression in all SCD patients with early AVN, and our results document that core decompression was effective for almost all patients with stage I or II AVN. Many of these patients were nearly pain-free after the coring procedure. In this series, 2 patients underwent core decompression when their disease was relatively advanced (stage III). With only 2 patients in this group it is not possible to determine if core decompression is beneficial in patients with advanced AVN. Our results, however, do concur with other series and indicate that stage III disease may be too late for intervention [14].

In our study, the patient's report of reduced pain and increased activity was chosen as the primary determinant of efficacy. It is interesting that although 10 of 11 patients with early disease had significant clinical improvement in pain and activity, 3 of 11 (27%) progressed on plain radiography. This discrepancy between clinical and radiographic findings underscores the need for uniform criteria to define efficacy.

Because of the morbidity associated with hip replacement and the historically poor results of hip replacement in SCD, the need for further surgery is an important secondary endpoint. Though the length of follow-up is relatively short, no patient treated with core decompression has required further surgical intervention. Core decompression is a much simpler procedure than hip arthroplasty and patients, generally, do not experience the more serious complications seen with total hip replacement. The only reported complication of core decompression has been intra- or perioperative fractures of the femur. Total hip replacement has a high rate of associated throm-

botic events, and patients often require a large number of transfusions. Sickle-cell disease patients are at particular risk of alloimmunization, and the Preoperative Transfusion study in SCD [16,22] documented increasing rates of alloimmunization as the number of units transfused increased.

Progression on X-ray was the third endpoint used to evaluate patients. The Cooperative Study of Sickle Cell Disease reported that radiographic findings in AVN were not a good indicator of clinical symptoms (2). Similarly, radiographic progression was often not associated with clinical failure in our patients, as 60% of those who had X-ray changes had significant improvement in pain and mobility. Progression on plain radiographs may be important, however, in long-term outcome, and extended follow-up of these patients is needed to answer this question.

All our SCD patients with early AVN underwent core decompression and, therefore, the expected outcome with conservative management alone cannot be determined. Natural history studies in SCD children with AVN documented that 80% remain symptomatic from diagnosis, and continue to have pain with follow-up as long as 18 years [1,2]. Despite the risks and high failure rate, almost one third of SCD patients with AVN will undergo hip replacement [1,2]. From these historical data it appears that the short period of bed rest after the coring procedure was probably not responsible for our patients' improvement, and supports our conclusion that core decompression improves the outcome of AVN in SCD. Due to the increased morbidity of surgery in SCD patients, however, a prospective, randomized trial is needed to better define the natural history of AVN in SCD, and to definitively recommend core decompression over more conservative therapies.

In summary, core decompression was effective in reducing pain and increasing mobility in SCD patients with stage I or II AVN. Symptomatic relief of pain often did not correlate with radiographic findings, as several patients had clinical success but progressed on radiography. Core decompression was associated with few complications, and no patient has required further surgery. However, due to the lack of natural history data and the risks of surgery in patients with SCD, a prospective, randomized trial is needed to definitively establish the benefit of core decompression in SCD patients with AVN.

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